

Short report

Paranganglioma of the cauda equina: a case report

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SUMMARY This case report describes a paranganglioma of the cauda equina in a 63 year old woman. Very few examples of paranganglioma have been recorded in this site. Histologically these neoplasms may have considerable similarity with ependymoma, the most common neoplasm of the lower spinal cord, and the diagnosis can be easily missed unless special techniques are employed. The clinical and pathological data obtained from this and the other reported examples suggests that parangangliomas of the cauda equina are benign, slowly growing neoplasms. In contrast to ependymomas and to parangangliomas elsewhere, they are well circumscribed, amenable to complete resection and have an excellent prognosis.

Parangangliomas are neoplasms that are thought to arise from cells containing catecholamines, which are widely distributed throughout the body in association with the autonomic nervous system.¹ Extra-adrenal tumours occur most often in the carotid body, glomus jugulare, mediastinum and para-aortic region. Parangangliomas from different sites show variable light microscopic patterns. An arrangement of compact cell nests is common, but the key diagnostic feature is the presence of cytoplasmic granules, only demonstrated reliably by electron microscopy.¹ These are electron dense, spherical, neurosecretory type granules, 100–200 nm diameter. The granules contain catecholamines, but in extra-adrenal sites the majority of these neoplasms are non-functional.

Case report

Clinical features

A 63 year old woman presented with a 12 year history of pain in her left buttock radiating to the posterior aspect of her left thigh. Initially she had ignored her symptoms which were intermittent, but 2 months prior to admission her pain became almost continuous and was exacerbated by exercise. Her walking distance progressively diminished and exercise

provoked a feeling of severe numbness in her left buttock and sacral area. She also began to experience a feeling of incomplete emptying of her bladder after micturition. Her general health was good.

On admission the only abnormal physical signs observed were in the skeletal and nervous systems. She showed a mild increase in the lumbar lordosis, associated with slight loss of flexion. There was a slight loss of sensation over the left buttock and sacral area and also the anterior aspect of the left leg. The left ankle jerk was moderately reduced. There was no significant loss of straight leg raising. Radiographs of the lumbo-sacral spine showed scalloping of the posterior aspect of the vertebral bodies with enlargement of the interpeduncular distances at the levels L1–S1. A myelogram demonstrated an intradural lesion occupying the lumbo-sacral canal from L2–S1.

Surgical procedure

When the dura was opened, after total laminectomy from L2 to S1, a dark red-brown tumour was exposed, extending from the level of L2 to S1. It appeared to arise from a lower nerve root on the left side and was firm and encapsulated. It required debulking with a Cavitron to aid dissection off the adherent nerve roots, before a complete removal could be performed. After operation she made an excellent recovery with greatly improved sacral sensation, but the left ankle jerk remained diminished. She did not receive post-operative radiotherapy. Twenty months later she is well with no evidence of tumour recurrence.

Histology

The resected tumour was light brown in colour, with a smooth surface and measured 7.0 × 3.0 × 2.0 cms. Histology revealed a densely cellular tumour composed of cuboidal to columnar cells with uniform rounded nuclei (fig 1). Mitotic figures were not apparent. In parts the

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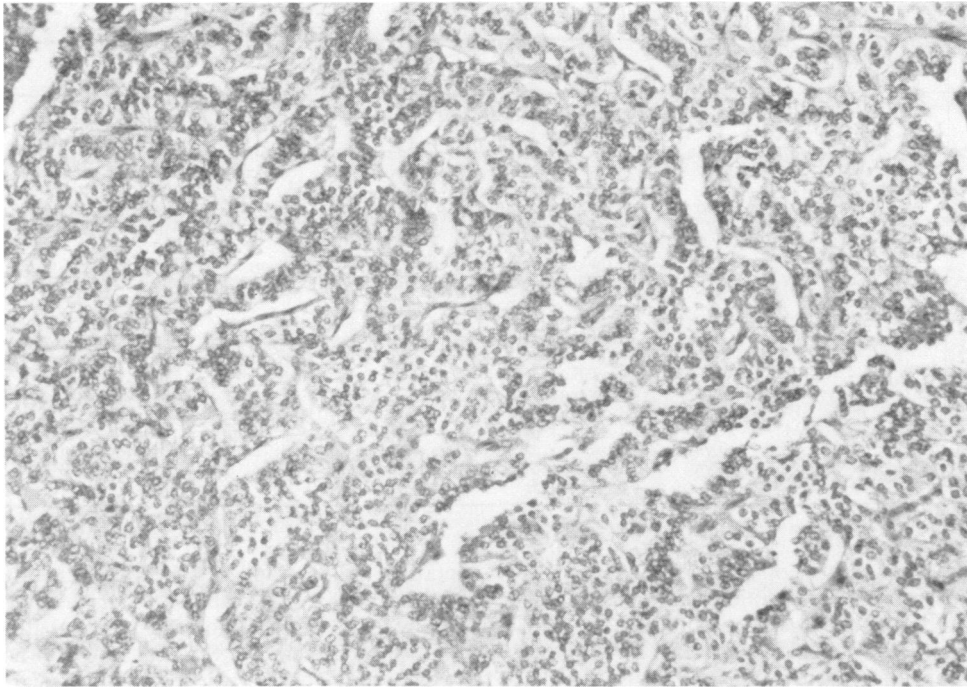


Fig 1 Tumour composed of cuboidal to columnar cells with uniform, small, rounded nuclei. Clefts and a ribbon-like pattern are apparent. A similar appearance may be found in ependymomas. (Haematoxylin and eosin $\times 130$.)

tumour cells showed clear cytoplasm and a packeted arrangement, elsewhere there was eosinophilic cytoplasm and a ribbon-like pattern (fig 1). There were occasional perivascular pseudorosettes, but no true rosettes. Inter-cellular stroma was scanty and inconspicuous throughout. A negative result was obtained with the immunoperoxidase method for glial fibrillary acidic protein. No mucin was demonstrable with either the periodic acid Schiff or alcian blue stain. The Grimelius silver staining method demonstrated fine cytoplasmic granules in many tumour cells.

Electron microscopy of formalin fixed tumour material revealed closely packed, interdigitating tumour cells with lucent cytoplasm. Small desmosomes were fairly common. There were occasional cells with dense cytoplasm. Numerous, small, rounded, membrane bound dense core granules, 100–200 nm diameter were evident in the cytoplasm of the majority of cells (fig 2). In addition many contained stacks of rough endoplasmic reticulum and in a few there were dense clumps of microfilaments. No microvilli or cilia were identified, although there were a few structures resembling ciliary basal bodies. These were in cytoplasm, unrelated to the cell membrane and were interpreted as centrioles. These findings established the diagnosis of paranglioma.

Discussion

The paucity of reported cases of paranglioma of the cauda equina suggest that either this is an extremely rare entity or one that has been frequently mis-

diagnosed. Probably both these statements are valid. This case emphasises the histological similarity with ependymoma, which is well documented as the most common neoplasm at this site.² It is important to be aware of the microscopic similarity so that appropriate special stains and electron microscopy can be employed to make the diagnosis. Accurate histo-

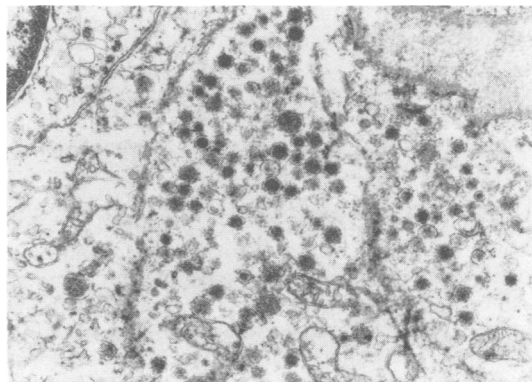


Fig 2 Numerous dense core granules in tumour cell cytoplasm. The granules have an outer membrane and dense central core, characteristic of catecholamine storing granules in normal paranglion cells. (Electron micrograph $\times 8\,000$.)

Table Data from reported cases of paraganglioma of the cauda equina

Author(s)	Age (yr)	Sex	Length of history	Clinical presentation	Size	Follow up
Miller & Torack ³	56	F	15 yr	Low back pain, leg weakness, urinary incontinence	26 gms	3 mo NRT, weak legs, incontinence urine and faeces
Lerman <i>et al</i> ⁴	29	M	13 mo	Low back pain, worse on exercise,	2 cms	18 mo NRT
Horoupian <i>et al</i> ⁵	59	F	1 yr	Low back pain worse on movement, retention of urine	1.5 cms	8 mo NRT neurogenic bladder, weak leg
van Alpen <i>et al</i> ⁶	44	M	6 mo	Low back pain	3.5 cms	2 mo NRT
Lagace <i>et al</i> ⁷	55	M	3 yr	Low back pain worse on exercise	2.5 cms	26 mo well, DXT post-op, removal incomplete
Llena <i>et al</i> ⁸	41	M	7 yr	Low back pain, acute urinary retention	8 cms	Fit 2 weeks post-op
Binkley <i>et al</i> ⁹	66	M	3 yr	Low back pain, weak legs, retention of urine	6 cm	8 mo NRT neurogenic bladder, weak leg
Llena <i>et al</i> ¹⁰	42	M	4 yr	Low back pain	3.5 cms	1 yr well NRT
Taxy ¹¹	52	M	5 yr	Low back pain, weak leg	Not stated	Incomplete removal, post-op DXT, recurrence after 9 years
	44	F	5 mo	Low back pain	3.5 cms	15 mo NRT
Soffer <i>et al</i> ¹²	13	M	2 mo	Low back pain	2 cms	Well post-op
Cabello & Ricoy ¹³	64	F	4 mo	Low back pain	2.5 cms	Not stated
	29	M	2 mo	Low back pain	2.7 cms	Not stated
Lipper & Decker ¹⁴	56	F	10 yr	Low back pain worse on exercise	3.5 cms	7 mo NRT slight weakness of foot
Anderson & Gullan (present case)	63	F	12 yr	Low back pain, incomplete emptying of bladder	7 cm	20 mo well NRT

NRT—no recurrence of tumour.

DXT—radiotherapy.

genetic classification of neoplasms is not just an academic exercise, but an important basis for selection of appropriate therapy, surgery, radiotherapy or adjuvant chemotherapy. It is important to segregate tumours which may have a different natural history, lest their inclusion in any clinical trial biases the results.

In this case, a long clinical history, radiological evidence of chronic bony changes and banal cytology suggest a benign, slowly growing, non-functioning neoplasm. Other reported cases lend support to this view. There is a fairly consistent record of a long history, often several years, of low back pain, prior to diagnosis (table). This is such a common clinical complaint that only exacerbation or the advent of additional neurological problems, such as leg weakness, bowel or bladder symptoms may prompt investigations. Urinary retention has been reported in several instances.^{5-8,9} In almost every case surgery has revealed a well circumscribed ovoid to cylindrical intradural, extramedullary tumour, with maximum dimension of a few centimetres, hence complete surgical resection has been possible (table). In a few cases it has been necessary to incise an adherent nerve root, but usually neural tissue has been preserved. In only one case, described as a secretory ependymoma, but probably the first reported example of a paraganglioma, was the filum terminale removed with the tumour.³ The local infiltrative growth, and spread to extradural soft tissue, that may occur with ependymoma, has not been described in paraganglioma at this site. Primary surgical excision appears to be the treatment of choice. As yet there has been no local recurrence after total macroscopic surgical removal (table). One case incompletely excised and given post-

operative radiotherapy recurred 9 years after the initial operation.¹¹ Another tumour that was only partially removed is said to have regressed after post-operative radiotherapy.⁷ Surgical resection has usually produced considerable relief of clinical symptoms. Mild neurological deficit has persisted in those patients where it was most severe preoperatively or where nerve roots were excised at operation. The available data suggest that paragangliomas of the cauda equina are slowly growing, well circumscribed neoplasms that may be cured by surgery alone. However, it is well known that paragangliomas in other locations, for example the retroperitoneum and larynx, have a poor prognosis, with high morbidity and eventual mortality,¹ hence there is a need for further documentation of the paragangliomas of the cauda equina, in order to establish whether or not their behaviour is uniformly benign.

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